# Hydatidosis: a global problem of increasing importance

R. M. MATOSSIAN,[1] M. D. RICKARD,[2] & J. D. SMYTH[3]

*This review of recent literature reporting the occurrence of hydatid disease due to* Echinococcus granulosus *and* E. multilocularis *throughout the world emphasizes the global nature of the problem and the threat of its spread into those countries currently free from it. Attention is drawn to the urgent need for measures to prevent the importation of infected livestock and this would require the development of techniques for pre-mortem diagnosis and differentiation of hydatidosis and cysticercosis of animals. There must also be increased awareness of the possible occurrence of biological strains of the parasite which may be of greater or lower infectivity for man. In the absence of information on infectivity, studies concerning the prevalence of the disease may be meaningless.*

It is well known that hydatidosis is a silent cyclozoonotic infection of cosmopolitan distribution and that larval forms of two closely related species of cestode, *Echinococcus granulosus* and *E. multilocularis*, are responsible for most of the overt cases of the disease, although other species with sylvatic cycles are known to exist. In man, the symptoms of unilocular hydatid disease are usually manifested several years after exposure. In animals, autopsy studies reveal an earlier invasion of cystic ·masses in the viscera. Multilocular (alveolar) hydatid disease has a more progressive course, characterized by an initial tumour-like process in the liver that subsequently metastasizes to the brain, lungs, and other organs. *E. granulosus* usually propagates through a simple domestic cycle, while *E. multilocularis* may go through a variety of wild animals that have different susceptibilities. Although the seriousness of the health hazard represented by hydatidosis has been reviewed frequently *(1–4)*, the appearance of the disease, within recent years, in countries previously free of it has produced an entirely new global situation. This appears to us to require urgent evaluation and forms the topic of this paper; emphasis has been placed on new records, and those areas that have already been adequately reviewed have been dealt with summarily (Fig. 1).

A comprehensive review of echinococcosis should consider acceptable methods for describing the regional variations that may occur in its distribution. The prevalence of adult worms in the definitive host, the presence of the larval stages in slaughtered ruminants, and the number of reported cases in humans have been used as criteria in assessing the prevalence of the infection. Gemmell *(1)* adopted a system whereby " An average incidence of 20% or more in one or more species of definitive or intermediate host was regarded as high ". Accordingly, the prevalence of the infection was considered as being high or low, widespread or restricted ón the basis of its periodic reported prevalence. Such a classification, though relatively biased, has served as a guide to indicate trends in the distribution of the disease. The use of serological screening in hydatid morbidity studies has often been hindered by the lack of sensitivity and specificity of the tests *(5)*.

## WORLD DISTRIBUTION

### A. *Iceland*

Throughout the 19th century Iceland had the highest prevalence of human hydatidosis ever recorded *(6, 7)*. However, prevalence rates of cysts discovered at autopsy declined from 25% (1900) to 16% (1932), 6% (1944), and 0% (1960). Sheep and dogs have also become free from infection *(7)*. This

[1] Professor of Bacteriology and Virology, American University of Beirut, Beirut, Lebanon.

[2] Senior Lecturer in Parasitology, University of Melbourne, School of Veterinary Science, Werribee, Victoria, Australia.

[3] Professor of Parasitology, Department of Zoology, Imperial College, London, England.
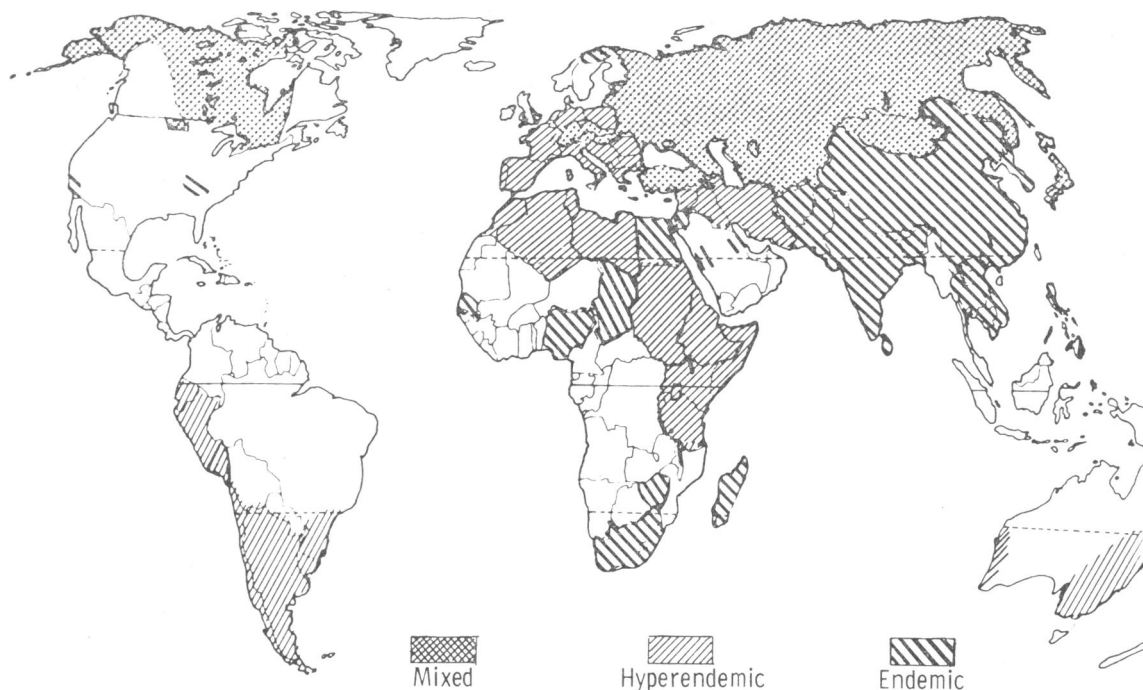
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Fig. 1. Global distribution of human and animal hydatidosis. Single lines represent *Echinococcus granulosus* and crossed lines the coexistence of both *E. granulosus* and *E. multilocularis* infections.

is ascribed to the strict application of prophylactic methods and to education of the public.

## B. *Europe*

Hydatidosis is a well-known phenomenon in Europe (*1–4*). A high prevalence belt of *E. granulosus* infection extends from the Iberian Peninsula to the Balkans. Reports from Bulgaria, Corsica, southern France, Italy, Greece, Portugal, Romania, Sicily, Spain, and Yugoslavia have stressed the hyperendemic nature of the disease (*1, 2*). Incidence rates are lower in Belgium and the northern parts of the Federal Republic of Germany (*3*) and in the Netherlands (*8*). A sylvatic cycle of *E. granulosus* infections, involving deer, dogs, and Kautokeino Lapps has been observed in northern Scandinavia (*9*). Equine hydatidosis is increasing in the United Kingdom (*10*), although a parallel rise in human cases has not been observed.

Infections with *E. granulosus* and *E. multilocularis* coexist in eastern Europe and extend all the way to the Pacific Ocean (*4*). A large zone of mixed infections also exists in central Europe (*4*). This includes Austria, the eastern regions of France, the southern districts of the Federal Republic of Germany, and Switzerland. Drolskammer et al. (*11*), in a review of 351 verified cases of echinococcosis observed in Switzerland (1956–69), described 64% as having unilocular and 35% multilocular cysts. Individual cases of alveolar disease have also been reported from Bulgaria, Greece, Poland, and Yugoslavia.

## C. *Asia*

### I. *South-West Asia and the Mediterranean*

The high prevalence belt of hydatidosis (*1*) prevailing in the Balkans spreads into Turkey and extends further into Iraq and Iran. These are primarily agricultural countries with the great mass of the population living in rural areas.

*Turkey.* *E. granulosus* infections are widely scattered in the country. A yearly average of 300 human cases has been reported by Oytun (*12*). The preva-

lence rates in sheep, cattle, goats, buffaloes, pigs, and camels have ranged from 40% to 70% (12, 13). There is an age-dependent increase in the number of cysts in slaughtered sheep and cattle, and 93% of sheep and 47.5% of cattle cysts were fertile (14). Fifty-two confirmed cases of E. multilocularis infection have so far been observed in Turkey (13), and adult cestodes were present in Turkish foxes.

*Iraq.* Hydatid infections constitute a major health problem in Iraq and their seriousness has been recognized by Senekji & Beattie (15), Babero et al. (16), Niazi (17), and others. In a country-wide study of the infection, Babero et al. (16) observed cysts in 29.5% of sheep, 13.9% of cows, 35.6% of buffaloes, and 49.1% of camels; of 169 street dogs captured in Baghdad and other cities, 38.4% harboured adult worms. Over 500 cases of human illness are recognized yearly with a wide variety of clinical manifestations (16, 17). No confirmed cases of human alveolar hydatidosis have been seen.

*Iran.* Hydatidosis, though endemic in Iran, is confined more to the northern and western provinces of the country (18). Human cases have been regularly observed in Teheran, Isfahan, and Shiraz although their yearly incidence is unknown. Estimates of cysts in slaughtered animals have varied; thus, in Isfahan, 6.6%, 4.5%, and 22.1% of sheep, goats, and cattle, respectively, were found infected (19), while in Teheran, 16.8% of sheep and 26.4% of cattle had cysts (20); of 955 camels examined, 54% were infected (21). Half of the street dogs in Isfahan and 3%–26.6% in Teheran were infected (22). In addition, 5% of jackals and 100% of wolves harboured adult worms. E. multilocularis was present in 10% of 30 red foxes (Vulpes vulpes) captured in the northern province near Azerbaijan (22), and two species of rodent, Microtus socialis and Allactaga elater, were found to have the larval stages of the parasite. Two human cases of alveolar echinococcosis were recently reported from the region (20).

*Syria.* Studies undertaken some 40 years ago by Turner et al. (23) demonstrated the endemic nature of hydatidosis in the country with 28.5%, 41.4%, and 27.8% of sheep slaughtered in Damascus, Homs, and Aleppo, respectively, having cysts. Syria allows the transit of sheep, goats, cattle, and camels from Turkey and Iraq to Lebanon, Jordan, and Saudi Arabia and these animals may spend several weeks there before they reach their destination. Syrians with hydatid disease have been treated in Lebanese hospitals.

*Lebanon.* Many reports concerning Lebanon have suggested the endemic nature of hydatidosis in the country (24). However, results should be interpreted carefully to differentiate imported infections from those acquired locally. The medical facilities available in Beirut have attracted patients from a wide area and therefore reported cases of hydatid disease may not necessarily be representative of Lebanon. The sheep and cattle slaughtered in Beirut abattoirs are imported from Syria, Iraq, and Turkey. Only pigs are bred locally and the high prevalence (31%) of cysts in them is indicative of the endemicity of the disease (25). Regardless of their origins, 23% of sheep and goats, 37.8% of cattle, 60% of donkeys, and 100% of camels were found to be infected (26) and their carcasses have produced infection in over 30% of stray dogs in Beirut (27). Epidemiological studies of human cases by Abou-Daoud & Schwabe (28, 29) revealed a 2:1 ratio of Christians to Muslims in Lebanon. This may reflect the reluctance of Muslims to keep dogs as household pets.

*Jordan.* The endemic nature of hydatidosis in Palestine was reported by Witenberg (30) and Torrance (31). These authors noted a high incidence of cysts in slaughtered animals and the common occurrence of human patients in Jerusalem, Jaffa, and other cities in the area. In a recent report of 12 cases of pulmonary hydatidosis, Sliman (32) confirmed the endemicity of the infection in Jordan.

*Israel.* Israel has had a high morbidity rate of about 100 cases per annum of human hydatidosis (5 per 100 000 population) (33). This, however, has been ascribed, in part, to the immigration of infected patients. Among slaughtered domestic animals, its prevalence has varied from 0.02% in Tel-Aviv to 12% in Beersheba. The clinical, surgical, and diagnostic problems of hydatidosis have been described by Levy (34), Peller et al. (35), and Lass et al. (36).

*Cyprus.* Cyprus has been considered as a high-prevalence island similar to Corsica, Sardinia, Sicily, and the Baleares, off the coast of Spain (1, 3, 37).

*Kuwait.* During the first half of the 20th century, the prevalence of hydatidosis in Kuwait was negligible. The first published report by El-Gazzar & McCreadie (38) described 51 patients of which only 5 were native Kuwaitis. A significant increase in human cases encouraged Hassounah & Behbehani (39) to study the epidemiology of the disease in the country. Infection rates in dogs captured in the cities and rural areas of Kuwait averaged 23%. Cysts were found in 32.5% and 40.2% of native and imported

Somalian cattle, while 11.4% and 10.4% of Iraqi and Saudi Arabian sheep were also infected. These findings indicate that Kuwait has become a high-prevalence area.

*Saudi Arabia.* Hydatid disease is not unknown in Saudi Arabia. Occasional cases have been observed in the eastern Province (*40*) and others were seen in the west coast (*41*). Infected sheep (10%) have been exported to Kuwait (*39*). At present, hydatid disease is not considered a major health problem in Saudi Arabia.

*Oman.* Oman is a vast country that is undergoing rapid development owing to its oil resources. There is no record of hydatidosis.

## II. *The Indian subcontinent*

*Afghanistan.* In a survey for echinococcal infections in Afghanistan, Buck et al. (*42*) collected data about dogs and slaughtered animals, and about human antibody levels. The presence of Echino-coccus-like ova in dogs, of hydatid cysts in ruminants, and elevated antibody titres in human adults led to the conclusion that hydatid disease represented the most important helminth health problem in the country.

*Pakistan.* Little is known about the prevalence and epidemiology of hydatid disease in Pakistan. Siddiqui & Siddiqui (*43*) described 24 cases of echino-coccosis seen during a period of two years in Jam-shoro, Hyderabad.

*Bangladesh.* Islam & Rahman (*44*) collected information about 18 human cases in Bangladesh. A significant number of cattle were also found to be infected. The authors concluded that echinococcosis was more common than hitherto appreciated.

*India.* The endemic nature of hydatidosis was reported from India by Maplestone (*45*) and Sami (*46*) and they described a high frequency of human and animal infections in south-western Punjab. 27% of dogs and 89% of cattle were found to be infected. In 1968, Reddy et al. (*47*) reviewed the 527 cases of hydatid disease observed in India. Though spread all over the country, the highest prevalence was in Andra Pradesh and Madras. Studies of a village in southern India revealed that 22% of the inhabitants were reactive to the intradermal test. One-third of street dogs and a high percentage of ruminants were also infected; 64.6% of cysts in sheep and 41.7% of cysts in cows were fertile. The worms were identified as being similar to the *Echinococcus granulosus granulosus* found in New Zealand (*48*). The endemicity

of hydatidosis in or around Delhi and New Delhi, in north-west India, was described by Prakash et al. (*49*). Devadason (*50*) concluded that echino-coccal infections should be taken into consideration by those who are planning the health and welfare of the country.

*Sri Lanka.* Native hydatidosis is considered to be a rare human condition in Sri Lanka (*51, 52*) and the few cases observed were in foreigners. However, a sylvatic cycle of transmission involving the jackal on the one hand, and herbivores such as the sambar (*Cervus unicolor*), deer, buffalo, and boar on the other, has been described. The infection may then pass to cattle and goats and to dogs.

## III. *South-East Asia*

*Peninsular Malaysia, Sabah, and Sarawak.* Hydatid disease is considered to be rare in this area. Duguid et al. (*53*) reported the first case of a locally acquired pulmonary infection in a child. Cysts have been observed in cattle, sheep, and pigs in Peninsular Malaysia (*54*).

*Indonesia.* The archipelago is another hydatid free area. Carney et al. (*54*) were first to report the presence of adult worms in two dogs captured in Sulawesi (Celebes) Island and the cystic forms of *E. granulosus* were seen in cattle, sheep, and pigs from Sumatra, Madura, and Bali.

*Democratic Kampuchea, Lao People's Democratic Republic, Thailand, and Viet Nam.* E. granulosus infections are widely spread in these countries, al-though the incidence rates in man are unknown. Larval forms of the parasite have been observed in humans, cattle, sheep, and pigs in Lao People's Democratic Republic, Democratic Kampuchea, and Viet Nam (*54, 55*).

## IV. *Eastern Asia*

*Japan.* Although the disease is uncommon, both forms of echinococcosis have been present in Japan. Yamashita (*56*) described 59 human cases of unilo-cular and 43 of multilocular hydatid infection. Cysts in sheep and pigs have occasionally been observed, although adult worms are rare in dogs. *E. multilo-cularis* infections in Japan are concentrated in the islands of Rebun and Hokkaido (*4*).

*The People's Republic of China.* Information about echinococcosis in China is relatively scanty. The endemicity of the infection was recognized in a review of the 27 articles published in Chinese during the period 1955–1964 dealing with case reports, sur-

gical treatment and diagnosis of human *E. granulosus* infections (*57*).

*Korea.* The endemicity of bovine echinococcosis (25% infected) is described by Issiki (*58*), and dogs are the definitive hosts.

## V. *Union of Soviet Socialist Republics*

Both forms of echinococcosis are endemic in the USSR. A vast literature (*4*), primarily in Russian, is available.

## D. *Africa*

### I. *North Africa*

*Algeria, Morocco, Tunisia.* These countries are primarily agricultural, rural areas where echinococcosis is hyperendemic. Chenebault (*59*) discussed hydatid disease in Morocco. During 1964, 200 cases were seen in Casablanca and in 1951, 23% of sheep, 43% of cattle, and 100% of camels were infected. In a similar study in Algeria, Pampiglione (*60*) concluded that hydatidosis was widespread in the country. Cherid & Nosny (*61*) described in detail the pathology and clinical manifestations of the disease in this area. Tunis is another high prevalence area; the figures for animals collected by Menchari (*62*) varied from 8%–86% for cattle, 7% for sheep, and 28% for pigs. Ben-Osman (*63*) estimated at about 600 the yearly number of human hydatid cases reported from Tunis.

*Libya.* Chest radiography of 42 167 Libyan Arab nomads revealed 147 patients with hydatid disease, later confirmed by surgery (*64*). This indicated a high prevalence of the disease in Libya.

*Egypt.* Though echinococcosis is endemic in Egypt, human infections are comparatively less common than in other Mediterranean countries (*65*). The prevalence rates in sheep, buffaloes, and cattle were 1.5%, 16%, and 10% respectively. Camels had a higher prevalence (31%) and a greater percentage of fertility (63%) of cysts (*66*). Of 573 stray dogs from Cairo, 4% had adult *E. granulosus* (*65*).

### II. *East Africa*

Human hydatidosis exists in several localized areas in East Africa while *E. granulosus* zoonoses are more widely scattered. The cycle is primarily the dog–ruminant–man type. In the Sudan, seroepidemiological surveys by Cahill et al. (*65*) support the impression that human and animal hydatid disease is endemic, more especially in the south. Fuller (*67*) conducted clinical and serological tests in the Dassanetch tribes of Ethiopia, and his results suggest an extremely high prevalence of hydatidosis in the area. In Kenya, more than 30% of the cattle, sheep, and goats have hydatid cysts (*68*). However, human hydatidosis is observed, mainly in Turkana, a desert region in the north of the country (*69, 70*). The main cycle of transmission appears to be between dogs and domestic livestock. Extensive studies of Nelson & Rausch (*71*) failed to demonstrate the existence of any sylvatic cycles in wild rodents. Although hydatid disease is present in cattle from all over the country, human cases have been observed only in the provinces of Uganda that are close to Turkana and southern Sudan (*72*). In Somalia, seroepidemiological studies, conducted in human and animal samples, have suggested that although the disease is endemic in animals, the prevalence in humans appears to be low (*73*). The presence of infection in 40% of Somali cattle exported to Kuwait supports this observation (*39*).

### III. *Central and West Africa*

Little information is available about this section of Africa. A report by Sirol & LeFevre (*74*) from Chad, in Central Africa, indicates the presence of the disease in humans and its endemicity in camels. In West Africa, the disease has been considered to be rare. Chabal et al. (*75*) reported the third case of human hydatidosis in Senegal, and Alabi & De La Cruz (*76*) reported a single case from Nigeria.

### IV. *Southern Africa*

Information is available about Southern Rhodesia and the Republic of South Africa only. According to Wolinagren et al. (*77*), echinococcosis is common in cattle but human infections are rare in Southern Rhodesia. Verster & Collins (*78*) studied the prevalence of hydatidosis in South Africa and found the infection in animals was widely scattered and its distribution had regional variations. Domestic dogs and wild jackals were disseminators and human infections were considered uncommon.

## E. *The Americas*

Echinococcosis in the Americas seems to be more prevalent in the north of North America and the south of South America. The endemicity of the infection in the north has been described by Rausch (*79*); the high-prevalence areas of South America have been reviewed by Gemmell (*1*), Williams et al. (*3*), and Schantz et al. (*80*). Less is known about the countries in Central America.

## I. North America

*Alaska.* Echinococcal infections are endemic. However, the strain of *E. granulosus* indigenous in this area has a primarily sylvatic cycle, deer being the intermediate hosts and wolves and dogs the definitive hosts. Domestic animals are not easily infected (*79, 81*). This strain may be responsible for the larger number of small pulmonary cysts per host and infections may often be asymptomatic. *E. multilocularis* infections of foxes (*Alopes* and *Vulpes*) and microtine rodents also occur in Alaska (*80*). Man may be infected following exposure to ova excreted by sledge dogs harbouring adult worms.

*Canada.* Infection with *E. granulosus* var. *canadensis* is endemic among the Indians, in the northwest, and the Esquimos of the eastern Arctic zone. The cystic forms of the parasite are found in the lungs of deer, but not in domestic herbivores, and dogs are the definitive hosts (*82*). Alveolar echinococcosis is widely spread throughout Canada (*3, 4*).

*The United States of America.* Though relatively uncommon, *Echinococcus* infections appear to be widely spread in the USA (*3*). Extensive epidemiological surveys have revealed foci of hydatidosis in Mississippi (*83*), California (*84*), and Utah (*85*). *E. multilocularis* adults were found in red foxes (*Vulpes vulpes*) and coyotes captured in North Dakota (*86*), and larval forms were present in 4.8% of deer, 4.8% of mice, and 1.9% of meadow voles trapped in the same area. Eearlier studies by Carney & Leiby (*87*) had revealed a similar presence of the parasite in Minnesota.

*Mexico.* Mexico appears to have the lowest incidence of hydatidosis in North America. The fifth autochthonous case was reported by Biagi & De La Garza (*88*). Results of seroepidemiological studies conducted in Oaxaca, Mexico, have indicated a low prevalence of reactors at diagnostic titres (*89*).

## II. South America

Southern South America includes some of the countries that have the highest prevalence of *E. granulosus* infection in the world. The magnitude of the problem has been reviewed by Williams et al. (*3*) and Schantz (*79*). Selected reports from Argentina (*90*), southern Brazil (*93*), Chile (*91*), and Uruguay (*92*) emphasize the hyperendemic nature of the disease in humans, domestic ruminants, and dogs. Peru is another country with high prevalence (*94*). In Colombia, though the disease is of low prevalence, the introduction of sheep from Argentina and

dogs from England may initiate another outbreak of hydatidosis (*3*). Thatcher (*95*) described 11 cases of echinococcosis in Colombia. Six of these were of the multilocular type. The author considers *Echinococcus oligarthrus* as the species responsible for the infection. Cysts recovered from spiny rats, pacas (*Cuniculus* spp.), and opossums were also considered to be *E. oligarthrus*. In Ecuador, the infection appears to have a low prevalence (*96*). Hydatid infections are rare in Venezuela. However, a case of multilocular illness possibly due to *E. multilocularis* was reported by Grases & Salazar (*97*).

## III. Central America

Though little is known about the nature of hydatid infections in this area, Williams et al. (*3*) consider the disease as being sporadic and confined to imported cases.

## F. Oceania

Australia (including Tasmania) and New Zealand have been considered as high-prevalence areas for hydatidosis (*1*). In Australia, the infection is prevalent in the southern part of the country (*98*) and in Western Australia (*99*). Following intensive programmes of control through education and public health measures, the disease is on the decline in New Zealand and Tasmania (*100*).

### DISCUSSION

It is apparent from this survey of the literature concerning the occurrence of hydatid disease in man, that since the publication of previous reviews on the subject there have been many reports of hydatid disease in previously free countries. Because most of the figures for prevalence in man rely on clinical cases, and because hydatid disease usually manifests itself only several years after infection, the data from these areas must underestimate the seriousness of the problem. Levels of infection in domesticated animals may more closely indicate the potential threat to public health. The appearance of hydatid disease in hitherto free areas could be the result of either better detection and reporting of the disease in the human population, or the recent importation of the disease into the country. It is this latter point to which we wish to draw particular attention.

Measures for the control of hydatid disease in endemic areas are well known, although often extremely difficult to implement. However, scant attention has always been paid to measures that might be

employed to prevent the extension of this disease into other countries. A major factor in the spread of *E. granulosus* must be the importation of infected livestock into those countries with developing agricultural industries. Although cognizance may be taken of more spectacular epidemic diseases of imported animals, hydatid disease is not considered. A major problem is that there is at present no reliable way in which this disease may be diagnosed in live animals in any domesticated species. Although serological diagnosis should be possible, the wide range of parasites, especially closely related organisms such as *Taenia ovis, T. hydatigena,* and *T. saginata,* with which the intermediate hosts may be infected makes specific diagnosis difficult. Despite this problem, we feel that the development of serological tests for hydatid disease in domesticated animals is an objective that must be pursued vigorously if control of dissemination of the disease is to be achieved. With hydatid disease the saying " prevention is better than cure " has real meaning; even in New Zealand and Tasmania, where control programmes have been in operation for two decades, total eradication has not been achieved.

Another factor that demands much closer attention is the possibility that there may be many biological variants or " strains " of *E. granulosus,* and that some of them may not be infective for man.

There is already evidence that such " strain " variation does occur. Smyth & Davies (*101*) reported that the " equine strain " and " ovine strain " of *E. granulosus* in the United Kingdom exhibit very considerable differences in development under comparable conditions of *in vitro* cultivation. Furthermore, Thompson & Smyth (*102, 103*) showed that despite an alarming increase in equine hydatidosis there had not been any evidence for an increase in the number of human cases. Also, Thompson & Smyth (*103*) were unable to infect rhesus monkeys (*Macaca mulatta*) either with eggs administered orally or by intraperitoneal injection of protoscolices of equine origin. If biological strains exist that are either not infective for man or are highly infective for man, then surveys of infection rates in the wide variety of reservoir hosts available may have little meaning.

In conclusion, in spite of several WHO/FAO-sponsored meetings on cysticercosis/hydatidosis held in recent years (e.g., Munich, 1974; Kenya, 1976), there does not appear to have been any increase in public awareness of the global nature of the disease and the threat of its spread into those countries currently free of it. Unless urgent action is taken to redress this situation, the natural pattern of the disease is likely to change for the worse in the next few years.

## ACKNOWLEDGEMENTS

## RÉSUMÉ

L'HYDATIDOSE: UN PROBLÈME MONDIAL D'IMPORTANCE CROISSANTE

La présente revue des publications récentes concernant l'apparition, dans le monde entier, de cas d'hydatidose dus à *Echinococcus granulosus* et à *E. multilocularis* met en évidence le caractère universel du problème et les risques de propagation de la maladie dans les pays qui en sont actuellement exempts. L'attention est appelée sur la nécessité de prendre d'urgence des mesures pour prévenir l'importation de bétail infecté, ce qui suppose la mise au point de techniques pour le diagnostic de l'hydatidose et de la ladrerie chez l'animal vivant. Il faudrait aussi se préoccuper davantage de l'existence possible de souches biologiques du parasite plus infectieuses ou moins infectieuses pour l'homme. S'il y en avait, leur infectivité à l'égard de l'homme devrait être clairement déterminée ou les études sur la prévalence de la maladie n'auraient guère de sens.

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